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Garcin syndrome: A rare complication of rhinocerebral mucormycosis in post covid patient

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ABSTRACT

Rhino cerebral mucormycosis (RCM) is now one of the rapidly emerging opportunistic, destructive and angioinvasive fungal infection with a very high mortality and morbidity. Early diagnosis of this condition is difficult as there are lacks of specific clinical features or manifestations. Garcin syndrome is more often caused by skull base and rhinopharyngeal tumors or metastases, and basal meningitis. Here we report an elderly female who was post covid status and presented with rhinocerebral mucormycosis and went on to develop garcin syndrome.

Keywords: Rhinocerebral mucormycosis, Garcin syndrome, COVID

1. INTRODUCTION

Mucormycosis is prevalent in nature but may be prone to cause infection in moist, temperate climates. Based on the anatomy mucormycosis is divided into six major forms which include rhinocerebral, pulmonary, cutaneous, gastrointestinal, disseminated and renal infection. Predisposing factors are uncontrolled diabetes mellitus, trauma, and corticosteroid use for prolonged periods, intravenous drug abuse, malnourishment and iron overload states. Most commonly diabetes mellitus is the underlying disease in patients developing mucomycosis, especially rhinocerebralmucomycosis (RCM) (Yang et al., 2016). Since there are no specific clinical features biopsy of the infected tissue revealing growth of the fungal agent is the gold standard. Thus, timely diagnosis of rhinocerebralmucormycosis is difficult. Garcin syndrome is characterized by progressive involvement of the cranial nerves which may lead to unilateral paralysis of all cranial nerves, no evidence of increased intracranial pressures, and skull X-rays showing osteoclastic lesions. Garcin syndrome along with rhinocerebralmucormycosis is a very rare condition (Nagendra et al., 2020).

2. CASE PRESENTATION

A 56 years old diabetic, non-hypertensive female presented with complaints of low grade fever, which was intermittent since 10 days. She also had complaints of cough since 10 days. She had complaints of swelling and redness of left eye since 5 days which was progressive. She also had complaints of headache since 5 days. Patient was recently diagnosed as COVID 19 one month back and had recovered from the same.

On examination, patient was afebrile with pulse rate of 136 beats/min regular, blood pressure was 100/80 mm of Hg in left arm supine position. Respiratory rate was 32/min with use of accessory muscles of respiration. Her Spo2 on ambient air was 84%. On examination patient had conjunctival congestion, chemosis and proptosis of left eye. CNS examination revealed intact higher mental functions. Cranial nerve examination revealed bilateral IIIrd and VIth cranial nerve palsy (figure 1 and 2); deviation of angle of mouth to right side and obliteration of left nasolabial fold suggestive of left VIIth cranial nerve palsy (figure 3). On asking the patient to say "ahh" uvula was deviated to right side indicating left Xth cranial nerve palsy and on testing gag reflex there was absent reflex in left side suggesting leftIXth nerve involvement. Respiratory sytem examination revealed bilateral crepts. Cardiovascular and abdominal examination was normal.

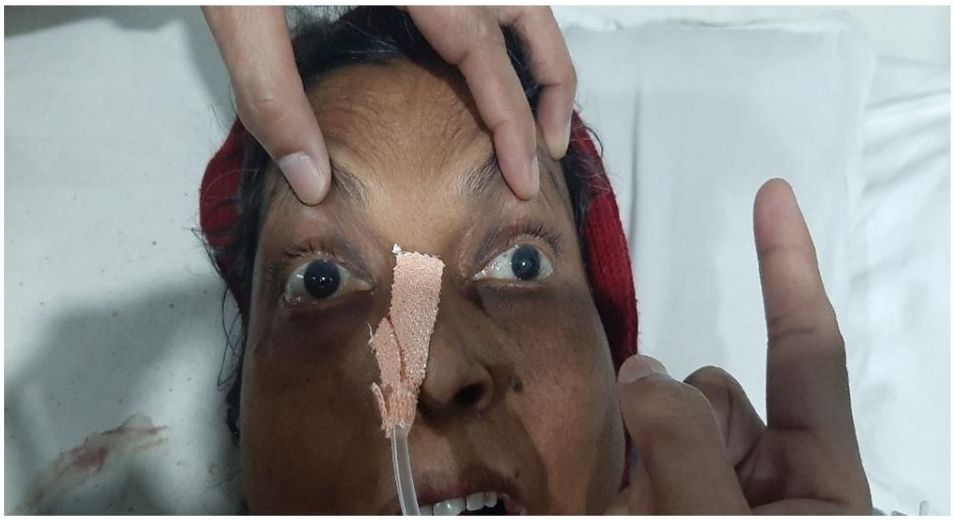

Figure 1 Examination of left eye revealing paralysis of left VIth and right IIIrd cranial nerve.

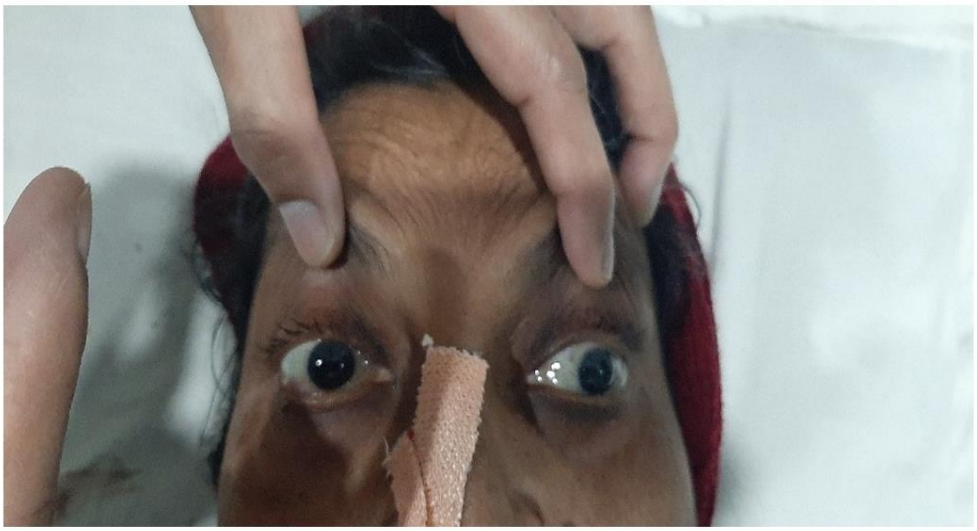

Figure 2 Examination of left eye revealing paralysis of right VIth and left IIIrd cranial nerve.

Figure 3 Examination revealed deviation of mouth to right side and obliteration of left nasolabial fold suggestive of left VIIth cranial nerve palsy.

Repeat reverse transcriptase polymerase chain reaction for COVID 19 was negative. Laboratory investigations revealed Hemoglobin 10.3 g/dL,total leukocyte counts 17,900/cumm andplatelet counts 2,83,000/cumm. Her PT INR was 1.12, PT was 13.4 (PT control was 12.5) and APTT was 30.8 (APTT control was 30). Her serum sodium was 135mg/dL, potassium was 4.4 mg/dL. Liver enzymes was normal (Aspartate aminotransferase of 16mg/dL and Alanine aminotransferase of 13mg/dl, total bilirubin 0.5mg/dL, conjugated bilirubin 0.2mg/dl, unconjugated bilirubin 0.3mg/dL, albumin 2.9g/dL, globulin 3.0g/dL). Serum creatinine was 5.6mg/dL, urea was 163mg/dL. Her CRP was 286mg/dL, LDH was 419mg/dL and ferritin 922mg/dL. Cerebrospinal fluid analysis revealed total leukocyte counts of 94 cells/cumm (Polymorphs 80% and lymphocytes 20%), pH of 7.2, glucose was 94mg/dL, protein was 288mg/dL, lactic dehydrogenase was 74mg/dL suggestive of tubercular meningitis.

HRCT thorax revealed multiple ill-defined patchy ground glass opacities with septal thickening and consolidation in bilateral lung fields suggestive of infective etiology possibility of atypical viral pneumonia, imaging grading was CORAD 6 and CT severity score of 14/25 (moderate). CT Paranasal sinuses revealed heterogenous density collection noted in bilateral maxillary, frontal, ethmoid and sphenoid sinuses predominantly on the right side with obliteration and widening of right sided maxillary ostium and causing mild erosion of the walls of ethmoidal air cells suggestive of sinusitis (Possibility of fungal etiology to be considered). The right orbit is displaced anteriorly suggestive of proptosis (figure 4 and 5).

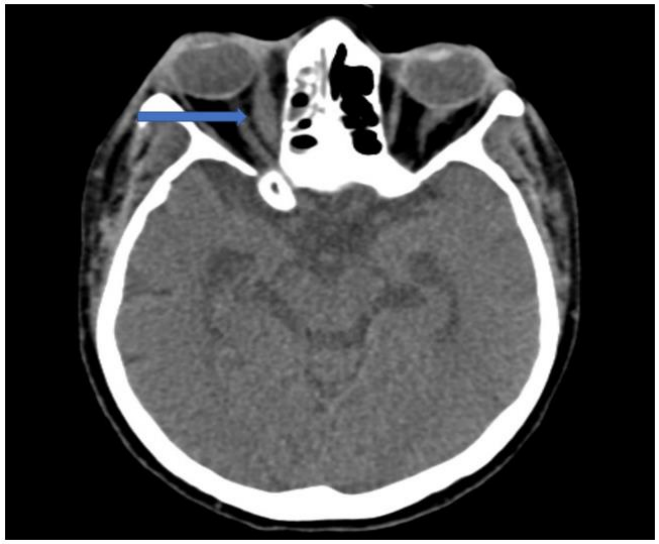

Figure 4 CT revealing blue arrow right sided proptosis with bulky medial recti muscle

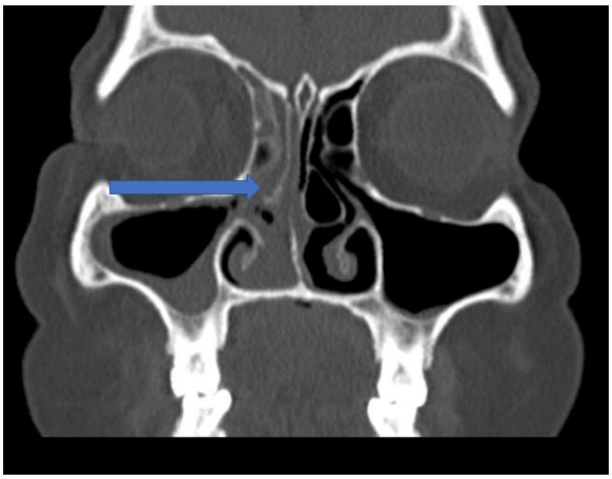

Figure 5 CT revealing blue arrow obliteration and widening of right sided maxillary ostium and infundibulum with extension to nasal cavity.

Patient was shifted to intensive care unit and immediately put on Bipap support (with a Fio2 of 100, Ipap of 18 and Epap of 12) and saturation improved to 90%. She was also started on liposomal amphotericin B, higher antibiotics and supportive management. Patient's condition gradually kept deteriorating; hypoxia worsened and patient's relatives were given option of invasive ventilation which they denied.There was increasing proptosis and blackish discoloration around theleft eye (figure 6).

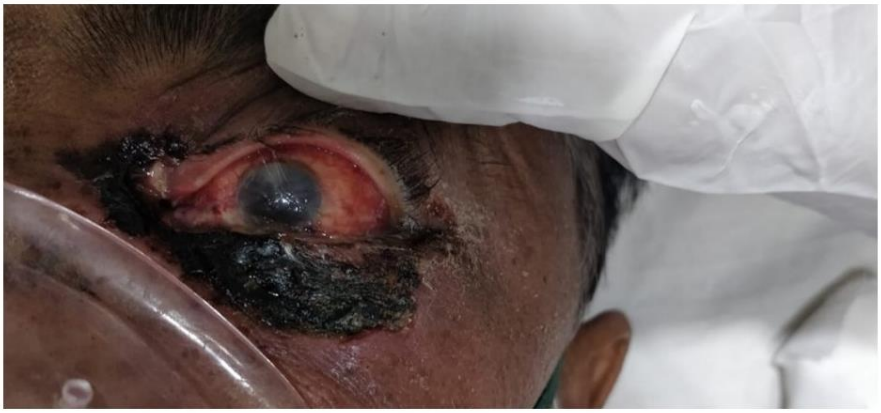

Figure 6 Proptosis and blackish discoloration around left eye.

As the patient was not able to maintain saturation and was on Bipap repeat CT scan could not be done. Opthalmology opinion was taken and urgent evisceration of left eye was done (figure 7 and 8). KOH mount of the tissue specimen revealed septate hyphae in chronic inflammatory background (figure 9 and 10). Post surgery patient's condition kept on deteriorating and was on continous Bipap support. Unfortunately 2 days later she succumbed.

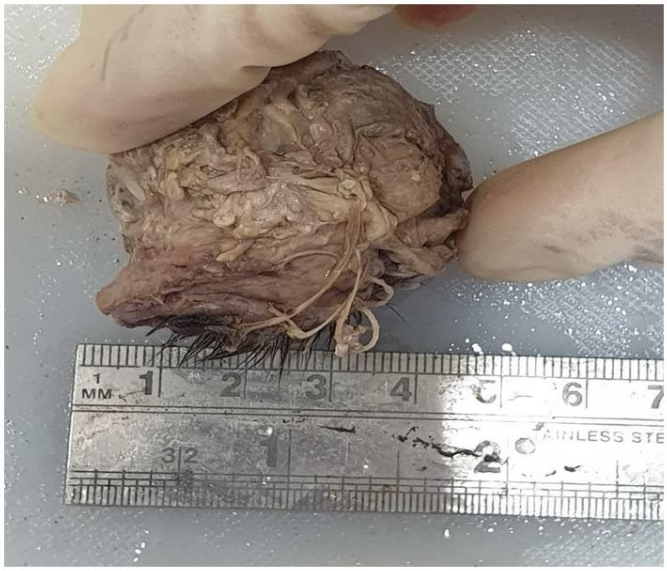

Figure 7 Specimen of the left eye which was eviscerated.

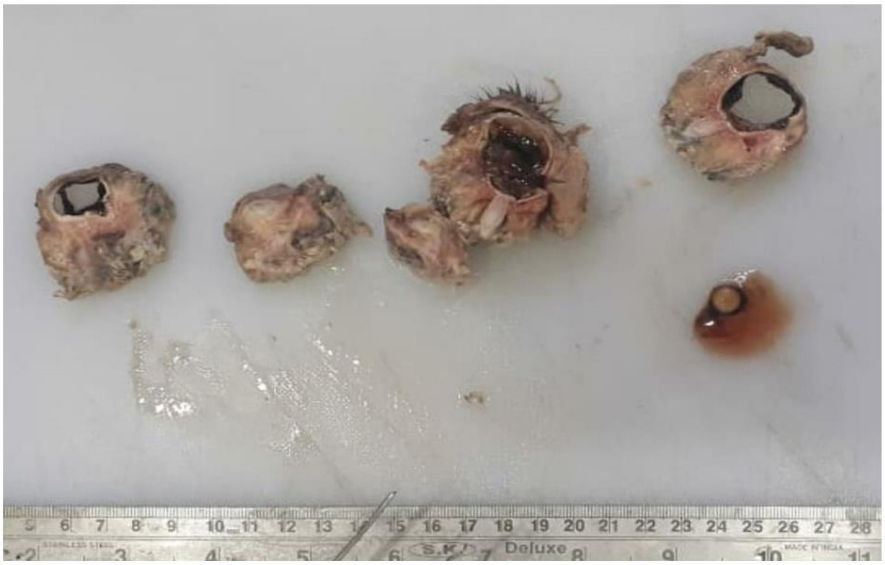

Figure 8 Serial transverse sectioning of eyeball reveals black discoloration of the vitreous humour, necrosis of the eyeball involving areas around the optic nerve as well

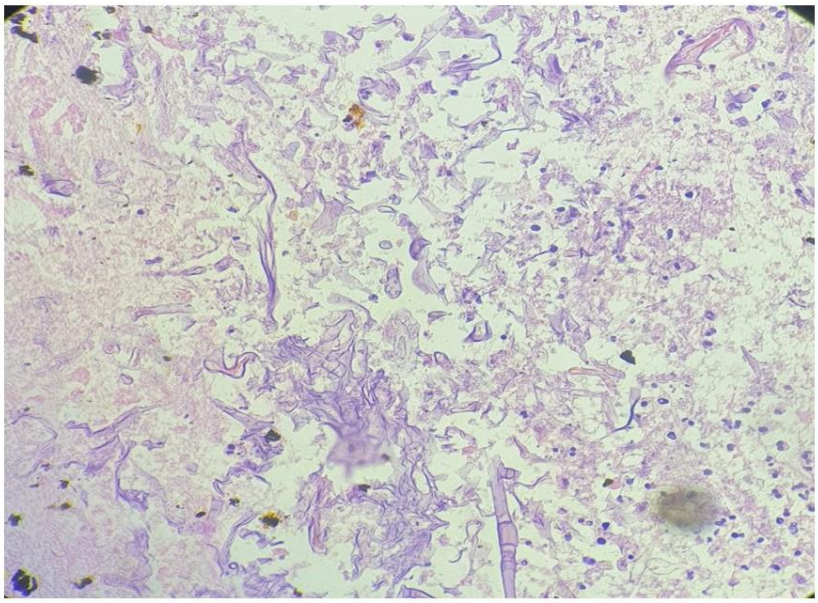

Figure 9 KOH mount showing broad, thick walled aseptate hyphae in a chronic inflammatory background

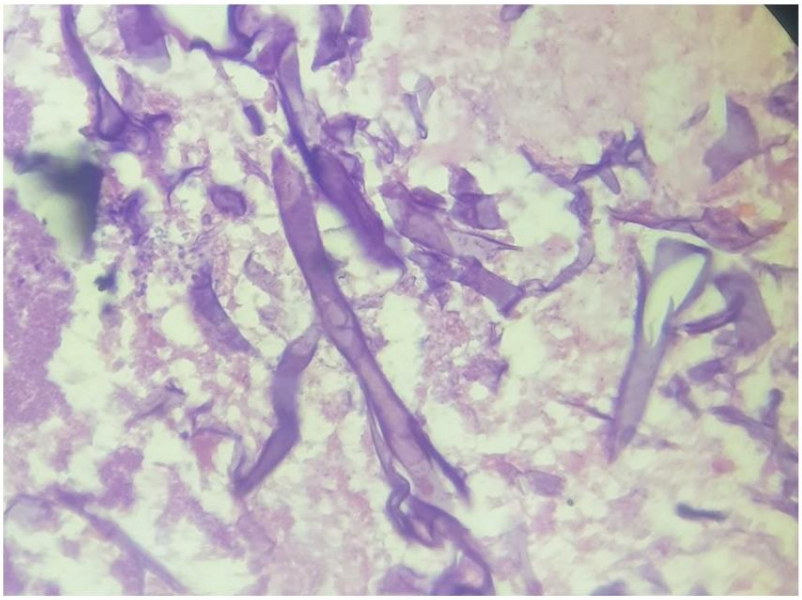

Figure 10 1000x magnification of non-septate, budding hyphae in a necrotic debri

3. DISCUSSION

Symptoms of rhinocerebral mucormycosis are non-specific complicating early diagnosis. The most common presentation includes fever, facial pain, headache, lethargy, visual loss, proptosis, orbital pain, palatal ulcer, black nasal secretions (Dronamraju et al., 2021). Rhinocerebral mucormycosis initially affects the paranasal sinuses and orbit and then spreads to the brain. There are complications of RCM like internal carotid artery and cavernous sinus thrombosis. Garcin syndrome is one of the rarest complications (Jain et al., 2011). Once the infection spreads into the cavernous sinus through local infiltration, IIIrd, IVth and VIth cranial nerve palsies occurs. In our patient cranial nerves IIIrd, VIth, VIIth, IXth and Xth are affected. Additional involvement of cranial nerves VII, IX and X could be due to local spread of infection in the neural tracts. This may lead to neural and perineural edema resulting in lower motor neuron type of cranial nerve palsy. Sarcoidosis and Wegener's disease also have a similar presentation and should be ruled out if patients don't have the associated systemic manifestations. Treatment of rhinocerebral mucormycosis includes administering systemic antifungals and performing urgent surgical debridement (Rajeshwari et al., 2012). Correcting hypoxia, acidosis, hyperglycemia and electrolyte abnormalities is critical for successfully managing such patients (Munir et al., 2007). In our patient despite starting aggressive antifungal therapy and evisceration of eye being done, the patient had an

unfavourable outcome. Underlying diabetes mellitus and poor renal function also contributed to poor outcome (Prakash et al., 2019).

4. CONCLUSION

In the current pandemic mucormycosis which is a life threatening condition is on the rise. It typically is affecting patients who are recovering from COVID-19 since they are immunocompromised due to COVID infection or due to the overzealous use of steroids and/or in patients who also have diabetes. Imaging, microscopy and histopathological analysis of biopsy specimens are used to confirm the clinical diagnosis. Repeated debridement may be needed and more severe cases may require orbital exenteration and removal of the sinuses. Simultaneously aggressive antifungal therapy should be started. Amphotericin B deoxycholate is the treatment of choice and liposomal preparations of this drug are preferred because of decreased nephrotoxicity. Posoconazole is the second drug of choice. Prognosis is poor even with aggressive surgery and anti-fungal therapy.

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Conflict of interest

The authors declare that there are no conflicts of interests.

Informed consent

Written and oral informed consent was obtained from the patient.

Data and materials availability

All data associated with this study are present in the paper.

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